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Cardiac Myxomas

Report of Eight Cases with Successful Excision

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Early diagnosis of atrial myxoma can be made by a high index of suspicion in patients in whom embolic phenomena, systemic manifestations, syncope, or suspected mitral or tricuspid valvular disease are noted. When the presence of an atrial myxoma is suspected, echocardiographic evaluation should be done promptly. This noninvasive technique has no morbidity or mortality and is accurate in greater than 90 percent of cases.

SINCE JANUARY 1964 excision has been carried out successfully in eight patients with cardiac myxoma at Loma Linda University Medical Center (LLUMC). Correct preoperative diagnosis was achieved in all, and there have been no recurrences. These cases are used as a basis for analyzing the modes of presentation and the course of right sided and left sided atrial myxomas.

Cardiac myxoma occurs with an incidence of 1/10,000 autopsies and the surgical survival in cases with correct preoperative diagnosis is substantially better (91 percent) than those in which the diagnosis was made at operation for suspected mitral valve disease (81 percent), as shown by a recent review of the past decade of surgical treatment of myxoma.[1]

Reports of Cases

Left Atrial Myxomas

CASE 1. A 9-year-old Puerto Rican girl was admitted to Loma Linda University Medical Center (LLUMC) on November 29, 1966, from a hospital in Puerto Rico.

Her first admission to hospital (in Puerto Rico) was in May 1966 for "probable rheumatic fever" with joint pains, nose bleeds, fever, fatigue and anorexia. The antistreptolysin O (ASO) titer was 1:625 and the erythrocyte sedimentation rate was 46 mm per minute. There was good response to treatment with penicillin, aspirin and steroids. In September 1966 she was readmitted because of an embolus to the left arm, which was removed surgically with good results. Her third admission to a Puerto Rican hospital was for the sudden onset of a severe headache, followed by pronounced lethargy, with receptive and expressive aphasia. After clinical resolution an angiocardiogram was done which suggested an intracavitary abnormality of the heart.

Physical examination on transfer to LLUMC on November 29, 1966, showed normal first and second heart sounds, a faint early diastolic sound, and a short midsystolic murmur at the lower left sternal border; one examiner heard a grade II/IV diastolic murmur at the apex. Small erythematous macules were noted on her left hand, with some cyanotic areas. Cineangiogram with pulmonary artery injection showed the presence of left atrial mass.

On December 1 the patient was taken to the

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operating room, where on cardiopulmonary bypass a friable gelatinous 3 by 4 cm tumor was removed. A broad pedicle was dissected away from the atrial septum where it was attached to the fossa ovalis. She was discharged in good condition eight days postoperatively.

CASE 2. A 31-year-old woman was admitted on June 20, 1968. She had had recurrent pulmonary infections over the past five years, and during the last 15 months had had progressive dyspnea on exertion, two pillow orthopnea, nocturnal dyspnea, pedal edema, fatiguability and a constant substernal pain. On physical examination a moderate right ventricular lift, a slight left ventricular lift and a palpable pulsation of the pulmonary artery were noted. The first sound and the pulmonary component of the second sound were both increased. There was a prominent early diastolic sound. A grade II/VI pansystolic murmur and a grade I/VI diastolic rumble were heard at the apex. On cardiac catheterization the pulmonary artery pressure was 34/14 mm of mercury and a pulmonary artery contrast media injection documented a filling defect in the left atrium on the angiogram. On June 28 the patient was taken to surgery, where, using cardiopulmonary bypass, a 5 cm globular myxoma, greenish tan in appearance, was removed. It was attached to the septum by a wide stalk, which was removed with a layer of atrial septal endocardium. She was discharged in good condition on the 12th postoperative day.

CASE 3. A 46-year-old woman was admitted on March 5, 1969, as a transfer from another hospital. (This case has been reported previously by Drs. Thompson and Simmons.[2])

During the first week of February 1969 the patient had had sudden onset of pain in the right eye, right side of the head and the left arm. On physical examination slurred speech and a cool pulseless left hand were noted. Results of cardiac examination were described on one occasion to be entirely normal except for a high pitched early diastolic sound. On another occasion a soft systolic murmur was heard at the left sternal border, which was not transmitted. Histologic examination of emboli removed from the right internal carotid and the left proximal brachial arteries showed tissue consistent with a cardiac myxoma. Cardiac catheterization and angiocardiography showed no abnormalities.

On March 17, 1969, the patient was taken to the operating room and cardiopulmonary bypass was established. At the junction of the posterior mitral valve leaflet and the posterior atrial wall there was an area about 14 mm in circumference, roughly round, where there appeared to have been friable tissue which had broken away. There remained a few friable projections, measuring up to 3 to 4 mm, and these were removed. Recovery was uneventful and she was discharged on the 13th postoperative day.

A repeat angiocardiogram was done in May 1974 because of bruits found on routine periodic physical examination by her private physician. Occlusion of the right internal carotid with cross filling from the anterior and middle cerebral systems was shown. The left axillary had occluded with distal reconstitution of brachial arteries to collaterals, with a possible false aneurysm forming just distal to the site of occlusion. She had no symptoms and no therapy was given.

CASE 4. A 54-year-old man was admitted on April 12, 1971, for exertional dyspnea, which had been present for a year and a half, paroxysmal nocturnal dyspnea six to eight months and occasional vague chest pain. On physical examination increased intensity of the first heart sound, a normal second heart sound and a faint intermittent early diastolic sound were noted. There was a grade II/VI middiastolic rumble with presystolic accentuation at the apex. A cardiovascular stress test (bicycle ergometer) on March 18 showed him to have normal exercise tolerance. He was able to exercise at 175 watts, and no signs of myocardial ischemia were seen on an electrocardiogram. Cardiac catheterization showed a systolic pulmonary artery pressure of 61 mm of mercury, and a large left atrial filling defect was shown on an angiocardiogram following pulmonary artery contrast media injection.

On April 22 the patient was taken to the operating room where, under cardiopulmonary bypass, a soft gelatinous gray-brown myxoma measuring 8 by 5 by 4 cm was removed. This was attached to the atrial septum in two places. The larger attachment was 2 cm above the mitral valve, and the secondary attachment was 1 cm higher on the septum. The pedicles and a layer of endocardium were removed, without creating a defect in the atrial septum. The postoperative course was uneventful and the patient was discharged on the eighth postoperative day.

CASE 5. A 67-year-old man was admitted on January 21, 1974, for evaluation of a calcified mass which appeared to be in the left atrium on

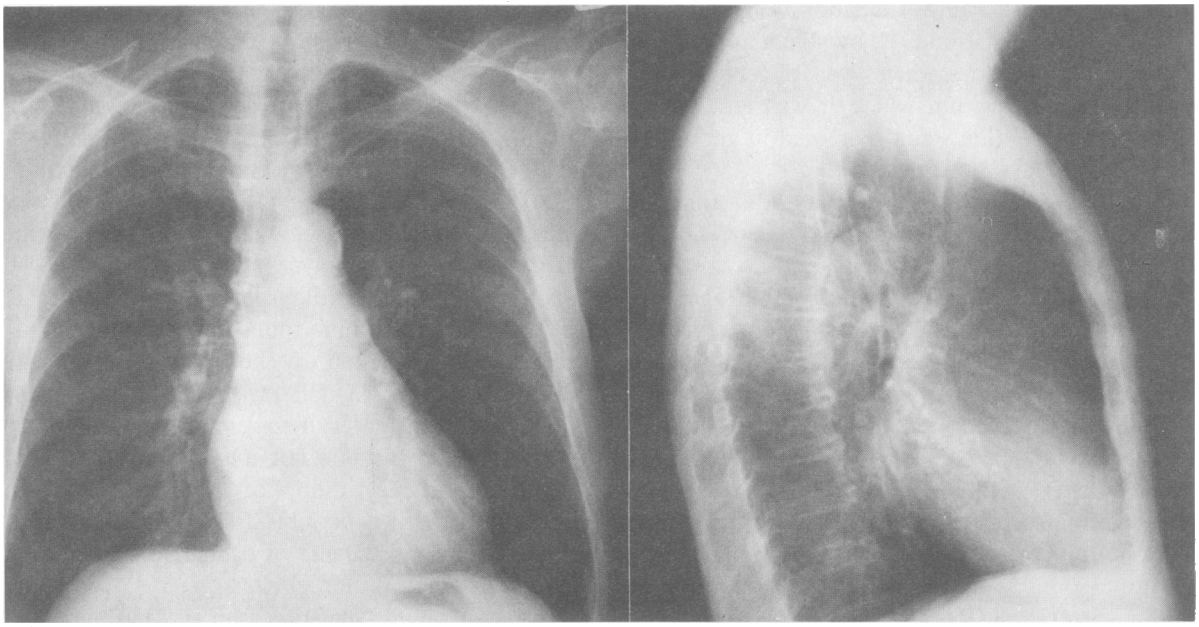

Figure 1.—Posteroanterior and lateral x-ray studies of the chest in case 5. Calcification is present in the region of the left atrium.

x-ray studies of the chest (see Figure 1) and follow-up tomography. He had been having episodes of dizziness associated with nausea, lasting about 20 minutes, and occurring two or three times a year for the ten years before admission. Symptoms had progressed during the five years before admission and included episodes of complete loss of consciousness and several grand mal seizures. On physical examination a totally irregular pulse with a rate of 120 per minute and a jugular venous pulse 4 cm above the sternal angle at 20 degrees were noted. A slight left ventricular heave was present. The first heart sound was split, and the second heart sound was normal. There were no murmurs. Cardiac catheterization with angiocardiography demonstrated a large left atrial filling defect. Echocardiography (see Figure 2) confirmed the presence of an atrial mass.

On February 22 the patient was taken to the operating room where on cardiopulmonary bypass a 5 by 5.4 by 4.5 cm grayish pink, nonfriable myxoma was removed (see Figure 3). Thrombus was present between the myxoma and the lateral part of the left atrial wall which partially enveloped a 3 by 4 cm area of the myxoma. The pedicle had a single attachment to the posterior atrial wall below the right inferior pulmonary vein. This was removed with a layer of endocardium. The postoperative course was uneventful although immediately after operation the patient had periods of confusion and disorientation. He was discharged on the 13th postoperative day. Atrial flutter was converted electrically to sinus rhythm several months after therapy with quinidine had failed to maintain sinus rhythm. Quinidine administration was discontinued because of thrombocytopenia. The patient was treated with prednisone for a short period with a good platelet response.

CASE 6. A 63-year-old woman was transferred from another hospital on March 5, 1974 because of multiple arterial emboli. She had had rheumatic fever at age five. A diagnosis of rheumatic heart disease with mild mitral insufficiency and possible minimal aortic stenosis had been made when the patient was seen as an outpatient five years before admission. Thirteen days before admission an acute inferior myocardial infarction had occurred. Embolism to the left subclavian artery occurred one day after the infarct, and to the right posterior tibial artery one day later. These were excised, but there was a poor response to the second embolectomy. The patient was transferred to Loma Linda University Medical Center where angiography showed an aortic saddle embolus and a right popliteal embolus. The day after admission the aortic saddle embolus was removed under general anesthesia. Findings on histologic examination of the embolus were consistent with car-

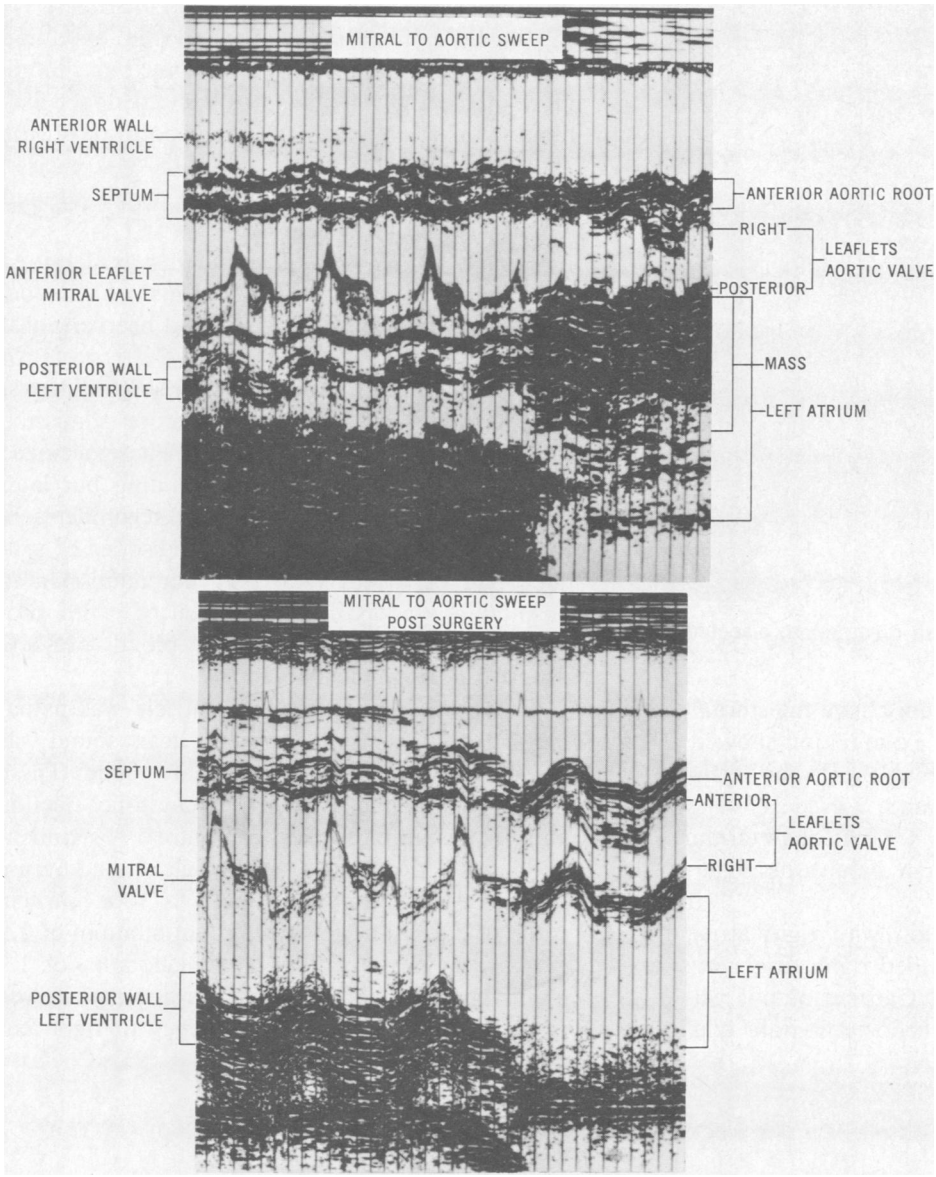

Figure 2. — Preoperative and postoperative echocardiograms, case 5. Dense mass in the left atrium before operation is absent in the postoperative sweep.

diac myxoma, and a left atrial myxoma was shown with angiocardiography. This myxoma could not be identified on an echocardiogram. The right leg became gangrenous and amputation below the knee was required.

On March 27 the patient was taken to the operating room, where on cardiopulmonary bypass a friable myxoma, 1.5 cm in diameter, which did not prolapse through the mitral valve was removed. A segment of the atrial septum at the base of the pedicle was removed, and the defect closed with sutures. There was no evidence of rheumatic valvular disease at operation. The postoperative course was complicated by a pleural effusion which was treated by thoracentesis, and the patient was discharged on the 21st postoperative day.

Right Atrial Myxoma

CASE 7. A 60-year-old man was admitted on August 17, 1970, for further evaluation of progressive dyspnea. Syncopal episodes had first occurred in December 1967. Right and left heart catheterization in June 1968 was not diagnostic. Because of increasing memory loss, weakness, weight loss, orthopnea and dyspnea, which was noted both on changing positon and on exertion after walking one block, reevaluation was done. On physical examination the patient was cachectic with an elevated jugular venous pulse with

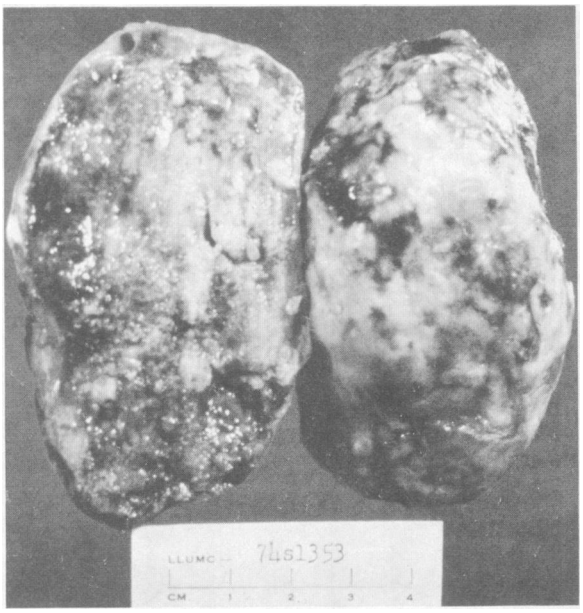

Figure 3.—Gross specimen of myxoma, bisected, from case 5.

prominent V waves, 8 cm above the sternal angle at 30 degrees. Cardiac examination showed a left parasternal heave and at the lower left sternal border a third heart sound, a grade I/VI systolic murmur and a grade II/VI diastolic murmur. The liver was palpable 6 cm below the right costal margin.

Cardiac catheterization with right atrial angiography showed a calcified right atrial mass, and tricuspid regurgitation. Cardiac output was 1.66 liters per minute. An echocardiogram confirmed the presence of a right atrial mass. On August 31, 1970, the patient was taken to the operating room where an 8 by 6 by 5 cm myxoma, filling most of the right atrium, was excised under cardiopulmonary bypass. A small portion of the atrial septum, near the fossa ovalis, was excised with the tumor pedicle and the defect sutured closed (See Table 1).

CASE 8. A 57-year-old woman was admitted to hospital on June 19, 1976, because of syncopal episodes. For several years she had been crippled by a spinal cord disease involving the posterior and lateral columns, but results on a Schilling test were normal. For one month before admission there had been syncopal episodes. These occurred chiefly when the patient was standing but later occurred when she was sitting or recumbent. In the two weeks before entry two episodes of syncope had occurred. Following her admission to hospital a syncopal episode occurred with pronounced hypotension. Systolic blood pressure dropped to 40 mm of mercury.

On examination the right ventricle was palpable in the epigastrium. The first heart sound was loud and there was a fourth heart sound. There was a low frequency harsh midsystolic ejection murmur in the right parasternal area. A sedimentation rate showed borderline elevation. Serum protein electrophoresis showed a total protein value of 5.5 grams per 100 ml, an albumin of 2.9 grams per 100 ml and a gammaglobulin of 1.1 grams per 100 ml. An electrocardiogram showed right atrial enlargement, a tendency to right axis

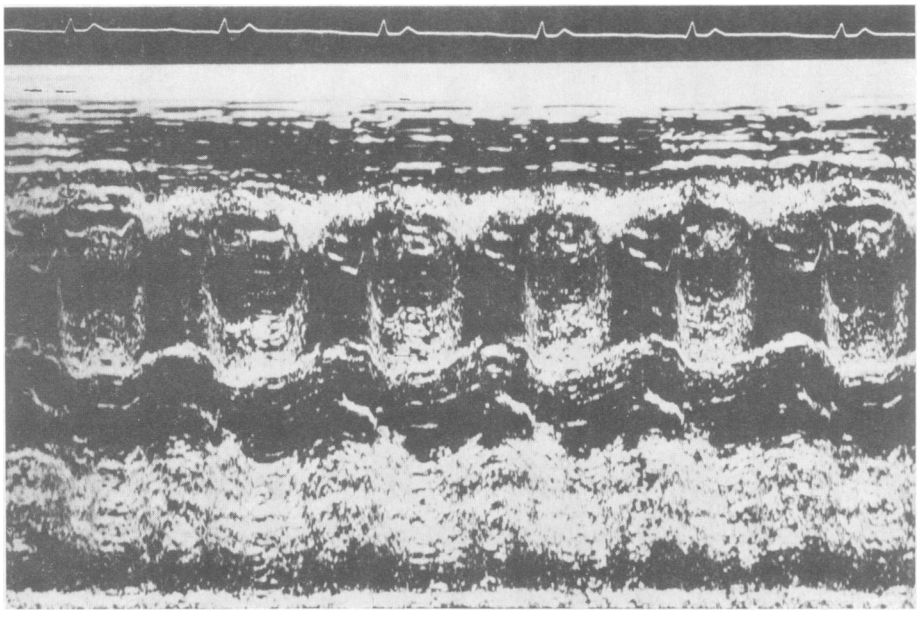

Figure 4. — Echocardiogram showing myxoma of right atrium.

TABLE 1.—*Summary of Clinical and Laboratory Features*

Left atrial myxomas—Cases #1-6

Pt. No./Age/Sex	Obstructive Symptoms	Emboli Location	Systemic Manifestations	Physical Findings	Laboratory						Catheter Data mmHg	
					Anemia	ESR	Serum Proteins Abnormal	Echo Cardiography	Ca^{+2} by X-Ray	EKG	PA	Wedge
1. 9 ♀	None	#1 Left brachial, #2 Cerebral	Fever, arthralgia, fatigue, anorexia	Faint EDS, short midsystolic M at LSB	−	+	+	ND	−	RVH	ND	ND
2. 31 ♀	DOE, orthopnea, PND, recurrent pneumonia	None	Weight loss, fever, chest pain, fatiguability	RV and LV heaves ↑S1, ↑P2 at apex	−	−	+	ND	−	Early RVH	85/50	20
3.* 46 ♀	None	Right internal carotid Left brachial	None	EDS, variable systolic M. at LSB	0	+	−	ND	−	Normal	25/5	10
4. 54 ♂	DOE PND	None	None	↑S1, EDS, DM at apex	+	+	+	ND	−	RVH LAH	61/23	31
5. 67 ♂	None	Episodic small cerebral emboli	None	Normal findings on cardiac examination	+	+	ND	+	+	Atrial fib.	35/18	18
6. 63 ♀	None	Coronary, left forearm, aortic bifurcation, right popliteal	None	↑S2, Grade II, SM apex→axilla	+	+	ND	−	−	Acute diaphragmatic infarct, antero-lateral ischemia	31/10	7
7. 60 ♂	DOE, orthopnea, PND, syncope	None	Weight loss, weakness	Prominent jugular V wave EDS, left parasternal heave, grade II DM	+	+	+	+	+	Atrial Fib. RBBB	"Normal"	"Normal"
8. 57 ♀	Syncope with ↓BP (systolic=40)	None	None	RV epigastric heave, ↑S1, harsh mid-systolic M Right para-sternal area	−	+	+	+	−	Right atrial enlargement, possible RVH	"Low"	ND

Ca^{+2}=calcification; DM=diastolic murmur; DOE=dyspnea on exertion; EDS=early diastolic sound; ESR=erythrocyte sedimentation rate; Fib.=fibrillation; LAH=left atrial hypertrophy; LSB=left sternal border; LV=left ventricular; M=murmur; ND=not done; PA=pulmonary artery; PND=paroxysmal nocturnal dyspnea; ↑P2=increased intensity of pulmonic second sound; RBBB=right bundle branch block; RV=right ventricular; RVH=right ventricular hypertrophy; ↑S1=increased intensity of first heart sound; ↑S2=increased intensity of second heart sound; SM=systolic murmur; Wedge=mean PA wedge.

*Previously reported.[2]

deviation and possible right ventricular hypertrophy. An echocardiogram (Figure 4) done on June 25 showed an abnormal shadow. The report indicated that "the shadow could, indeed, be due to a mobile tumor in the right atrium lodging in the tricuspid valve during diastole." On right heart catheterization the pulmonary artery and right ventricular pressures were both low. The right atrial mean pressure was 10 mm of mercury. A single contrast injection at the orifice of the superior vena cava showed a large tumor mass within the right atrium which prolapsed through the tricuspid orifice during diastole and swung back towards the right atrium during ventricular systole.

On June 29 a 7.5 by 5.5 by 4.5 cm yellow tan friable mass attached to the fossa ovalis of the atrial septum was surgically removed under cardiopulmonary bypass. The myxoma was removed with a 1 cm margin and a 2 by 3 cm woven dacron patch was sutured into the surgical defect in the atrial septum. At the time of operation a pacemaker was implanted. The patient made an uncomplicated recovery from the surgical procedures but still remained crippled from neurological disease. She was discharged on July 14 (see Table 1).

Comment

Crafoord in 1954 was the first to successfully excise a cardiac myxoma.[3] Angiocardiography was first used in the diagnosis of myxoma in 1950,[4] and echocardiography in 1968.[5] As more cases are being reported, a better idea of the clinical features has become available.

Left Atrial Myxoma

The clinical features of cardiac myxomas are related to systemic manifestations, hemodynamic obstruction or embolic phenomena.

MacGregor first described the systemic manifestations of weight loss, fever, anemia, increased sedimentation rate and elevated globulins.[6] Our first case was in this category, but was exceptional in the degree to which it mimicked acute rheumatic fever. Joint pains, elevated ASO titer, and epistaxis in a 9-year-old patient must be caused very rarely by myxoma, because no similar cases were encountered in a recent comprehensive review of the literature of myxomas treated surgically.[1] Cobbs alludes to personal experience with a myxoma case in which the symptoms were those of rheumatic fever, but he does not give details.[7] A myxoma has been reported in conjunction with rheumatic heart disease, but there was no evidence of rheumatic involvement in our patient at operation. Prominent systemic manifestations were also present in Cases 2 and 4.

The hemodynamic manifestations mimic mitral or tricupsid valve disease, depending on the location of the myxoma. In cases 2 and 4 there were left atrial myxomas with mitral murmurs, and notably elevated pulmonary artery pressures (85 and 61 mm of mercury, respectively).

Embolic phenomena occurred in cases 1, 3, 5 and 6, or two thirds of the left atrial myxomas in our series. This is an incidence of embolization comparable to the 59 percent recently reported for left atrial myxomas.[1] Suspected coronary artery embolism with acute myocardial infarction, as occurred in case 6, is very unusual, and has only been reported twice in the operative myxoma literature of the past decade.[8,9] The diagnosis of atrial myxoma was made by histologic examination of embolic material in three of our cases (1, 3 and 6).

Extensive calcification of myxoma which allows the diagnosis to be made on chest film or fluoroscopy was present in our patient with right atrial myxoma (case 4) and in one of our patients with left atrial myxomas. Calcification is far more common in right atrial myxomas (28 percent) than in left (8 percent).[1]

False negative results of echocardiography in our case 6 can be attributed to the fact that a large portion of the myxoma had embolized to various arterial sites including the aortic bifurcation. The remaining portion of the myxoma was small and did not prolapse through the mitral valve. The echocardiographic technique used by Sun to visualize nonprolapsing tumors,[10] which is sometimes technically very difficult, was not employed. Echocardiography is generally an accurate test for myxoma, and was positive in 49 of the last 54 cases in which surgical operation was done.

The pedicle usually arises from the atrial septum, near the margin of the fossa ovalis. This is used as one of the criteria for distinguishing myxoma from thrombus,[11] and is the basis for some of the theories of pathogenesis of myxoma.[12] Case 5 is an exception to this rule; in that case the pedicle arose from the posterior atrial wall below the right inferior pulmonary vein. In case 4 there were two pedicles, both arising from the atrial septum.

With the advent of echocardiography, there is now available a noninvasive procedure for evaluation in patients with mitral valve disease, syncope or suspected embolic phenomena. In case 5 there were normal findings on physical examination of the heart in the presence of a 5 by 5 cm myxoma. Symptoms had been present for ten years. In case 2 symptoms had been present for five years and were attributed to mitral valve disease. The patient in case 4 went to cardiac catheterization two years before the diagnosis was made at a subsequent study. During the past decade, 23 percent of the patients operated on for cardiac myxomas went to surgery with a presumed or cardiac catheterization "confirmed" diagnosis of mitral valve disease.[1] Earlier diagnosis, based on a high index of suspicion, and the more widespread use of echocardiography in the future should reduce the morbidity and mortality in groups such as these.

Right Atrial Myxoma

Characteristic hemodynamically obstructive elements were present in both of our cases of right atrial myxoma. Isolated tricuspid stenosis was present clinically in one case and this is very suggestive of right atrial myxoma.

Both patients also had syncope, which is more common in myxomas than in rheumatic valvular lesions.

Calcification had developed in the myxoma (in case 7) to the point that it could be recognized at fluoroscopy and with plain films by the time of the first heart catheterization, two years before. Calcification is far more common in right atrial myxomas (28 percent) than in left (8 percent).[1]

Results on echocardiography were positive, but (in case 7) the studies were done after angiocardiography had shown the presence of the tumor. Echocardiography is reportedly less useful in evaluating right sided heart tumors.[1] The tricuspid landmarks are more difficult to visualize than mitral landmarks, and in patients with large right sided tumors the anatomy of the heart may be distorted and tumors can be missed, or only suggested by ultrasound techniques.

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